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RECOGNITION AND TREATMENT OF CONGESTIVE HEART FAILURE IN INFANCY: II. TREATMENT*†

The treatment of infants suffering from the conditions discussed in Part I (January 1959), is governed by three major considerations: First, the prognosis with medical treatment alone is grave, however great the skill and tireless the devotion of the attending physician and however intelligent the parents. The exceptions are the self-limiting conditions of myocarditis, paroxysmal tachycardia, iatrogenic failure from overtransfusion and congestive failure due to coarctation of the aorta, which if once adequately controlled may never recur. Second, congestive failure and respiratory infection are very frequently present together. Third, surgery is available for most of these conditions. If Table I (in Part I) is re-examined, all the conditions marked with a single asterisk are operable and indeed for them surgery is imperative. The conditions marked with a dagger are potentially operable, and have been successfully operated upon, but the decision in an individual case remains difficult because the mortality is still fairly high. Surgery should not be denied on grounds of youth, nor because the infant shows temporary improvement with medical care. It is by no means justified to assume that this improvement will be maintained.

Treatment thus divides itself into three stages. These do not consist, as is frequently thought, of digitalis, oxygen and hope. The three stages are: (1) control of the acute episode of failure by digitalis or other means; (2) precise diagnosis, using appropriate special studies; and (3) surgery. If surgery is either impossible or not indicated, then long-term medical treatment is all that remains.

The time relationships here are important. Stage 2, i.e., precise diagnosis, should follow Stage 1 within a matter of days and certainly within a month. Many infants are lost each year because Stage 2 is bypassed and long-term medical therapy is embarked upon without precise knowledge of the underlying malformation. Later, when the infant again suffers a relapse, he is referred to a cardiac center and "last ditch" surgery is embarked upon. But now surgery is attempted in an infant who has been severely ill for several months, whose renal function may have been weakened by low salt intake and diuretics, whose lungs are filled with edema fluid and opaque with atelectasis or pneumonia. He may be likened to the parable of the man with the seven devils whose last state was worse than his first. Quite naturally the infant does not tolerate surgical intervention in this state.

STAGE 1—MEDICAL TREATMENT OF ACUTE EPISODE

The salient principles of rest, oxygen, digitalis and diuretics are applicable in infancy as in later life. *Rest* is the most difficult to ensure and the most neglected aspect of care, particularly in teaching hospitals. There is much anxiety and restlessness associated with congestive failure in infants, and often prolonged, irritable crying which is unrelieved by a pacifier, feeding or walking motion. Sedation should be employed either prior to or immediately following the initial examination and maintained at an adequate level. This usually necessitates the use of phenobarbital in 8 mg. or larger doses four hourly before feedings. A night's sleep often can be assured by Nembutal suppositories. The physical comfort of the infant is often overlooked. As Bonham Carter has emphasized, many of these infants are orthopneic and are more comfortable when nursed in a sitting position or on an inclined plane. Frequent vomiting and excessive sweating makes these babies a serious nursing problem. Much discomfort can be avoided by

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† Part I of this article, concerned with the diagnosis of congestive heart failure in infancy, appeared in the January issue.

Table I

Rapid Digitalization in Infancy

Cedilanid—Digalen

Timing	Route of Administration	Dosage in		Proportion of TDD Given
		µg./Kg. mg./Kg.	≡ mg./lb.	
Stat.	Electrocardiogram Cedilanid I. V.	20 µg./Kg.	0.01	½
6 hours	Electrocardiogram Cedilanid I. V.	8 µg./Kg.	0.004	¾
12 hours	Electrocardiogram Cedilanid I. V. Digalen I. M.	8 µg./Kg. 8 mg./Kg.	0.004 4.0	TDD Cedilanid ¼ Digalen
24 hours	Electrocardiogram Digalen I. M.	16 mg./Kg.	8.0	¾
36 hours	Electrocardiogram Digalen Oral	8 mg./Kg.	4.0	TDD Digalen
48 hours and then q. d.	Digalen Oral	3 mg./Kg.	1.5	—

Total digitalizing dose (TDD) of Cedilanid: 30-40 µg./Kg. (≡ 0.015 mg./lb.)
Total digitalizing dose (TDD) of Digalen: 30-35 mg./Kg. (≡ 15 mg./lb.)
Maintenance dose of Digalen about 1/10 TDD: 3 mg./Kg. (≡ 1.5 mg./lb.)

timing drug dosage, examinations, etc., so that the infant is not incessantly disturbed.

Digitalis

Excellent discussions on the use of digitalis in infancy are available in the textbooks of Taussig, Nadas, or Keith and there have been many recent articles on the subject. The preparations in current use are Cedilanid, Digoxin, digitoxin and Digalen. If the infant is acutely ill, one of the rapidly acting preparations must be given first, followed by Digalen or digitoxin. Digoxin has the advantage that if given intravenously it acts almost as rapidly as Cedilanid and can therefore be used for both stages of the digitalizing procedure.

Rapid Digitalization: This is indicated within 24 hours or less in very severe failure, especially left-sided, in paroxysmal tachycardia and in postoperative failure. It always requires hospitalization, since frequent electrocardiograms are essential (Tables I and II). Great caution must be exercised if the above scheme is used, since each infant has an individual tolerance for digitalis and in some this outlined course of therapy will prove too slow, while in others it is toxic and far too rapid. Danger signs are the sudden onset of vomiting, the appearance of bradycardia (apex beat 90 or less in a young infant) and most frequent of all, arrhythmias. When an electrocardiogram is taken during digitalization, it is remarkable how often it shows premature contractions, the Wenckebach phenomenon or auricular tachycardia with intraventricular block, which indicate quite clearly that toxic levels have been reached, yet the original plan is rigidly followed as if it were a magic formula. In *newborn infants* and in *myocarditis* toxic reactions are particularly common and it is wise, in most cases, to use a lower total digitalizing dose than outlined above and to proceed with great caution.

Table II

Rapid Digitalization in Infancy

Digoxin

Timing	Route of Administration	Dosage in		Proportion of TDD Given
		μg./Kg.	≡ mg./lb.	
Stat.	Electrocardiogram Digoxin I. V.	36	0.018	½
6 hours	Electrocardiogram Digoxin I. V.	18	0.009	¾
12 hours	Electrocardiogram Digoxin I. V.	18	0.009	TDD
24 hours	Electrocardiogram Digoxin Oral	12	—	—
36 hours and q. 12 hrs.	Digoxin Oral	12	—	—

Total digitalizing dose (TDD) of Digoxin: 65-75 μg./Kg. (≡ 0.035 mg./lb.)
Maintenance dose ¼-½, usually ⅓, of TDD: 15-25 μg./Kg. (≡ 0.010 mg./lb.)

Regular Digitalization: This may be accomplished satisfactorily by oral medication. Digalen can be used, the total digitalizing dose being 30 to 35 mg. per Kg., given in six divided doses over two to three days; or Digoxin, 60 to 75 μg. per Kg., given in six divided doses over the same period.

Maintenance Digitalis: Digalen, 2.5 to 3 mg. per Kg., once daily, is adequate in most cases; if Digoxin is given, the dose is 20 to 30 μg. per Kg., given in two divided doses per day.

Oxygen

Any infant undergoing rapid digitalization should be kept in oxygen. A cool moist atmosphere should be maintained and the infant's distress should not be worsened by overheating or by the use of a small, cage-like box.

Diuretics

These are extremely useful as a temporary measure to relieve acute failure, particularly pulmonary edema. However, mercurials are more toxic to the infant than the adult kidney, and long-term diuretic therapy is usually both dangerous and unsatisfactory. In most instances, except when acute fulminating pulmonary edema is present, it is wiser to wait until the cardiac output has been improved by digitalis and give mercurials the day after digitalization is complete. *Thiomerin*, 0.25 to 0.50 cc., intramuscularly or subcutaneously, will usually produce a good diuresis. It should not be repeated more often than every other day, and then only if frequent electrolyte studies can be made. *Diuril* appears useful as an auxiliary diuretic. We have been using a conservative dose of between 5 and 15 mg. per Kg. per day, given in two divided doses. Hypokalemia should be watched for with great care. Although in adults this appears to be a very safe drug, we do not yet know enough about its long-term effects in infants to use it on such a basis except in inoperable cases. *Lanolac* or low-sodium milk is useful in the first few days, especially if frank edema be present. The combined use of Lanolac and diuretics, however, requires extreme caution, since it easily produces hyponatremia; frequent electrolyte studies are essential.

Antibiotics

During the acute episode, antibiotics should always be given in full therapeutic doses. Penicillin alone or penicillin with streptomycin may

be used until the results of nose and throat cultures tell whether any other specific therapeutic agent is indicated.

ANCILLARY METHODS OF TREATMENT

Acute Pulmonary Edema

This may be seen in sudden, severe left ventricular failure due to coarctation, aortic stenosis, occasionally endocardial fibroelastosis or following surgery for severe pulmonic stenosis. *Morphine* is useful in this situation in a dose of 1 mg. per 5 Kg. and can be repeated every four hours, if necessary. Morphine is otherwise very rarely indicated and is of course contraindicated in severe right-sided failure, especially if due to primary pulmonary hypertension. *Aminophylline* may be used in this group of infants and in the treatment of bronchiolitis associated with heart failure. It is now recognized to be a toxic drug and fatalities have followed its use. Nevertheless, it may be lifesaving on rare occasions. *Venesection* is rarely employed, probably being most useful if overtransfusion is the cause of the edema. It may be replaced by the use of *tourniquets* on three limbs simultaneously and rotated at ten-minute intervals. One great advantage of tourniquet treatment is that it ensures the constant presence of the physician during its use and at the same time keeps him sufficiently employed! The successful treatment of an attack of pulmonary edema in a small infant is one of the most gratifying moments in pediatric cardiology.

Severe Anemia

This may complicate the picture of cardiac failure and may be treated, once the infant is digitalized, with small repeated *transfusions* of packed cells given very slowly under sedation and using a fifteen-minute pulse chart.

Effusions

Whether pericardial, pleural or ascitic, they may be relieved by *tapping*. Except for large pericardial effusions the infant is seldom embarrassed by large quantities of edema fluid and dramatic improvement seldom follows. Pericardial taps should be carried out under electrocardiographic control, with an electrocardiograph electrode attached to the exploring needle so that a current of injury can be immediately recorded if the myocardium is touched. Prolonged searching for small collections of fluid is exhausting and dangerous for the infant and should be avoided.

Paroxysmal Tachycardia

This disturbance has been well discussed by Nadas and recently by Husson. Rapid digitalization should be employed. The only room for controversy is when and if other drugs should be employed if digitalis fails. An infant will very rarely die from a paroxysmal tachycardia of less than three days' duration, so if he is seen early, digitalis should be given a full trial; up to one-and-a-half times the usual digitalizing dose should be given in 24 to 36 hours. However, in a late case, methacholine, in a dose of 0.20 to 0.30 mg. per Kg. subcutaneously, may be given or quinidine, 3 to 5 mg. per lb. Methacholine should be given, as emphasized by Husson, only under full electrocardiographic control and with an intravenous infusion running already so that atropine can be injected immediately to counteract any untoward reactions.

STAGE 2—ESTABLISHMENT OF THE PRECISE DIAGNOSIS

As previously mentioned, most cases can be assigned to one of the five large diagnostic groups without any special studies. Immediately the acute episode of failure is controlled, special studies can be undertaken.

Left-to-Right Shunt

In this group, cardiac catheterization is, as a rule, the most useful test, provided the infant is neither in such profound congestive failure or so anemic that the changes in oxygen content are obscured. Catheterization will confirm the presence of an atrial or ventricular shunt, or the catheter may pass through a patent ductus. If there is doubt clinically between a ventricular defect and a persistent A-V canal, catheterization should be accompanied by selective angiocardiography. *Aortography* is indicated if one is in doubt about the presence of an extracardiac shunt or one wishes to differentiate between a ductus and a truncus or aortic pulmonary window. *Angiocardiography* is particularly useful in confirming the presence of anomaly of the venous return into the left superior vena cava and is more pathognomonic and probably less hazardous than cardiac catheterization in that particular condition. In the other left-to-right shunts, angiocardiography is not precisely helpful unless it is selective in character.

Obstruction to Ventricular Outflow

Special studies are rarely necessary in pulmonic stenosis and catheterization may be hazardous. Left-heart catheterization should be done if there is doubt about the presence of severe aortic stenosis. The presence and position of a coarctation may be confirmed by aortography, but this is seldom essential.

Obstruction to Pulmonary Venous Return

Such obstruction is suggested by catheterization, which shows a high pulmonary artery and

pulmonary capillary pressure with no shunt. Angiocardiography is useful in localizing the site of the obstruction.

Transposition

The diagnosis is usually quite clear on clinical grounds. However, in case of doubt, angiocardiography is the simplest method of demonstrating the origin of the aorta from the right ventricle.

Myocardial Group

Angiocardiography has recently been used to differentiate the large dilated, thin-walled left ventricle of anomalous left coronary artery from the large hypertrophied thick-walled left ventricle of endocardial fibroelastosis. Aortography may confirm that the right coronary artery alone arises from the aorta. Catheterization is of use only to exclude a shunt.

STAGE 3—DECISION REGARDING SURGERY

This is too complicated and intricate a subject to discuss in the short space remaining. In general, operation should always be undertaken for patent ductus, pulmonary stenosis or anomalous pulmonary venous return once failure has occurred. In coarctation, long-term medical therapy may be successful and surgery should be undertaken only if adequate medical treatment fails. Cardiopneumonopexy is probably worth attempting in all cases of anomalous left coronary artery. It is not yet clear whether this operation should be combined with ligation of the abnormal vessel. The rationale for cardiopneumonopexy is less clear-cut in endocardial fibroelastosis. Great variations in prognosis render evaluation more difficult.

Operations for transposition carry a very high mortality in infancy and there is much room for discussion in each individual case as to whether the hazards are justified.

Ventricular septal defect has passed in the last few years from an inoperable to an operable condition. It is, however, still considerably more dangerous to operate in infancy than in childhood. Unless the infant is being referred to a center where operations on older patients are going extremely well, he will probably stand a better chance of survival with medical care.

LONG-TERM MEDICAL CARE

If surgery is contraindicated, impossible or postponed, then long-term medical care must be undertaken. Digitalis should be continued for a minimum of six months from the initial episode of failure, and more often should be maintained until two years of age. The dose will need to be increased slightly from time to time as the infant gains weight. A low-sodium intake can be maintained by the use of a half-and-half mixture of Lanolac and evaporated milk. Unless the infant is under very close and frequent supervision, Lanolac should not be given alone over long periods of time. Hyponatremia is a very grave complication in infancy. Diuretics should be given as rarely as possible. Prophylactic antibiotics, either oral penicillin 600,000 units daily, monthly injections of Bicillin, or Terramycin 10 mg. per lb. daily, are frequently of help in the prevention of the recurrent infections to which these infants are subject. The parents should be clearly warned of the hazards of infection. They should forbid handling of the infant by relatives with colds or coughs and should wear masks if they develop infections. It is frequently and tragically the case that the parents come to understand this problem better than the admitting officer of the hospital. During infancy, any respiratory infection accompanied by a fever of over 101 degrees, any signs of bronchitis or increased respiratory distress, require hospitalization and oxygen. Far too many such infants are sent home because they "do not have pneumonia," only to return the next day, or a few days later, almost moribund. The failing heart may cease to beat with an infection scarcely noticed in a normal infant. It is often wise to let the parents carry a note at all times which explains the infant's past history and precarious state, so he will not be turned away by an inexperienced house officer.

The parents need much comfort and reassurance in handling these infants. It is very hard to be both truthful and optimistic. One can with all conscience, however, assure the parents that with each succeeding month the infant's chances improve, and if he can be got to his first birthday he is past the overwhelming dangers of infancy and may well begin to surprise them by his improvement. They should realize he will grow slowly if he has a large left-to-right shunt and should not torment themselves by comparisons with "ideal" infants, or even those next door. Many parents are concerned to know if the infant will die suddenly at home. Some do, but it is rare, and in any case unavoidable, and it is kinder to tell them this will not occur so that the mother may have courage occasionally to leave the child. Adequate sedation of the infant is a necessary part of the management. Many need phenobarbital every four hours and almost all need a nightly sedative for many months.

Depending on the underlying diagnosis, x-rays and electrocardiograms should be repeated every one to three months. Repeated fluoroscopy is not indicated; a large heart will remain large, and when it does not and the infant is going to recover, then the long-term dangers of repeated fluoroscopy become important.

Those who have read the delightful book "Please Don't Eat the Daisies" will remember the incident of the headless stalks decorating the

dining room table. Similarly, in pediatric cardiology, one is not always cognizant of the errors that can be committed. One recent example may be cited: a little colored infant, making an excellent recovery from myocarditis, was sent home on an adequate dose of Digalen. However, the mother was advised to count the pulse before each dose and if the pulse rate was below 100, to omit the dose! She returned three weeks later maintaining that the rate had always been below 100 and so no digitalis had been given; the rate was actually 240 and the infant was in profound failure. Matters such as digitalis dosage are the physician's responsibility and must never be shifted to the parents. Once the maintenance dose is known for the particular infant, the mother can be told to omit one dose if vomiting occurs; if it recurs after the next dose, the infant should be seen immediately and readmission is often indicated. A vomiting baby in borderline failure requires very close attention.

Similar instances seen recently include the sudden changing from digitoxin once daily in adequate dosage to Digoxin once daily in inadequate dosage, either in ignorance of or oblivion to their differing excretion rates; or the unrelenting use of Thiomerin and Lanolac in the presence of hyponatremia and hypochloremia.

Despite our ignorance of many important facets of the recognition and treatment of congestive failure in infancy, it is one of the most fascinating and rewarding subjects in cardiology and there is no limit to its future growth.

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